Complex Vascular Ring Diagnosed on Cardiovascular MR in a 3-Day-Old Infant

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Abbreviations: Magnetic resonance imaging (MRI), computed tomography (CT)

ABSTRACT

Prenatal ultrasonography in the early third trimester showed an unusual branching pattern of the right aortic arch. Echocardiography performed 4 h after birth showed the right aortic arch with mirror-image branching, patent ductus arteriosus, and patent foramen ovale. Because the location of the ductus arteriosus was unclear on echocardiography, cardiovascular magnetic resonance imaging was performed 3 days after birth. Advanced techniques including contrast-enhanced time-resolved magnetic resonance angiography and 3D time-of-flight magnetic resonance angiography allowed accurate diagnosis of a vascular ring comprising ascending and descending aorta, right aortic arch with mirror-image branching, and diverticulum of Kommerell giving rise to a left ligamentum arteriosum. The infant had hiccups, but no other symptoms. The esophagram was negative for obstruction. The infant was closely monitored; however, she developed esophageal obstruction at 7 months of age because of the vascular ring. She underwent lysis of the left ligamentum arteriosum followed by aortopexy for relief of esophageal obstruction. This report shows the utility of *neonatal* cardiovascular magnetic resonance imaging to evaluate complex congenital aortic arch anomalies.

INTRODUCTION

A female neonate was delivered vaginally in the 37th week of pregnancy with no labor complications at our institution; her APGAR scores were 8 at 1 minute and 9 at 5 minutes. The neonate weighed 2,320 g and was deemed small for her gestational age. The mother has a history of Hodgkin lymphoma, which was diagnosed and treated with chemotherapy in 1997. In addition, the mother has a history of chronic hypertension for which she takes labetalol; she suffered 4 miscarriages; and she was anemic on admission for delivery. The pregnancy was complicated by intrauterine growth restriction and the presence of a two-vessel (single umbilical artery) cord. Following delivery, the neonate was mildly hypoglycemic, but the blood sugar levels normalized within 4 hours of delivery.

A vascular ring and ventricular hypertrophy were suspected on screening prenatal ultrasonography. Accordingly, renal ultrasonography and echocardiography were performed. The renal ultrasonography showed an ectopic left kidney in the lower abdomen. The initial echocardiogram performed within 4 hours of birth showed several cardiovascular abnormalities. Color Doppler showed a patent foramen ovale with bidirectional shunting. There was also tricuspid regurgitation and mild enlargement of the right ventricle with heavy trabeculation of the wall.

The echocardiogram also showed a right aortic arch with mirror-image branching, a left brachiocephalic artery, a right common carotid artery, and right subclavian artery branches, from left to right. A patent ductus arteriosus was present, with bidirectional, but mostly left-to-right, shunting shown on color Doppler. There were no other abnormalities in size, appearance, or presence of vasculature. Subsequent echocardiography performed 1 and 2 days after birth confirmed the presence of these anomalies, although the ductus arteriosus was not seen on either of the follow-up echocardiograms. Possible outpouching of the right ventricular free wall was additionally noted, although ventricular aneurysm versus absent pericardium was not definitively concluded from the echocardiograms (Figure 1).

A cardiovascular MRI using gadolinium contrast and computerized hemodynamic analysis was performed 3 days after birth using a 1.5 Tesla Siemens Aera scanner (Muenchen, Germany). The infant was swaddled, and no sedation was necessary. Cine balanced steady-state free precession sequences (trueFISP) in standard cardiac projections were acquired, along with double-inversion recovery sequence and phase-contrast imaging. Contrast-enhanced, time-resolved (TWIST) and 3D time-of-flight magnetic resonance angiography techniques were also acquired in the coronal projection. The MRI confirmed the presence of a patent foramen ovale with a bidirectional flow.

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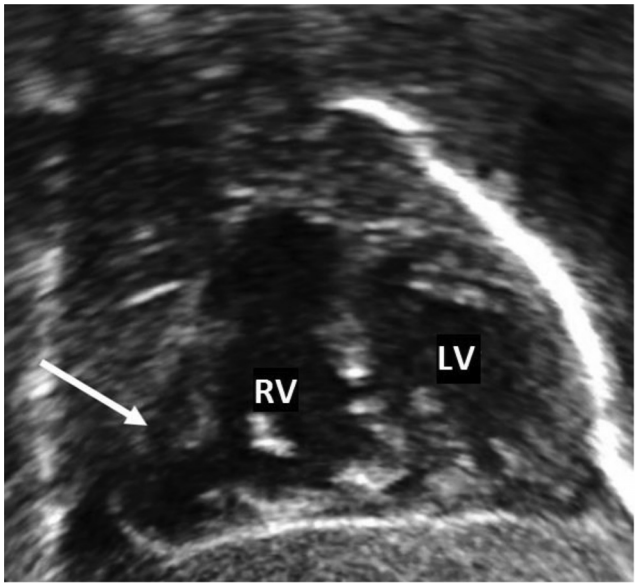

Figure 1. 2D conventional transthoracic echocardiogram showing focal outpouching (arrow) of the right ventricle at the acute angle of the heart.

The right atrial appendage was enlarged and extended along the free wall of the right ventricle. Cardiac MRI also showed increased trabeculations of the right ventricle and identified an outpouching of the right ventricle inlet at the angle of the heart, measuring 10.9 × 9.5 mm, compatible with a diverticulum. Biventricular cavity size and systolic function were normal.

Cardiac MRI also confirmed the presence of a right aortic arch with mirror-image branching (Figure 2, A–B). A diverticulum of Kommerell was present at the aortic isthmus measuring 7.1 × 4.4 mm, extending posterior to the esophagus (Figure 2C). A left-sided ligamentum arteriosum was also present, with interval spontaneous closure of the ductus arteriosus, completing the vascular ring (Figure 3, A–B). There was abutment but no obstruction of the adjacent esophagus and trachea by the vascular ring.

Esophagram performed during the first week of life showed ring displacement and mild narrowing of the esophagus but no significant impingement upon the trachea. At 7 months of age, the child developed dysphagia to thick liquids. She was taken to the operating room and posterolateral thoracotomy was performed. The vascular ring was identified and the left ligamentum arteriosum was seen tracking across the esophagus. The right arch was dissected and the ligamentum arteriosum was ligated and divided doubly. Aortopexy was also performed, relieving the esophageal obstruction. She was discharged a week later and remains well.

DISCUSSION

This neonate has a right aortic arch, mirror-image branching, a diverticulum of Kommerell, and a left-sided ligamentum arteriosum creating a complete vascular ring. The presence of a right aortic arch occurs in ~0.1% of the population and is due to improper regression of the embryonic aortic arches in the developing fetus (1-3). A right aortic arch forms following the failure of the right fourth embryonic aortic arch to regress along with abnormal regression of the left fourth aortic arch (2). The branching pattern of a right aortic arch is variable. An aberrant left subclavian artery pattern is most common, coursing posterior to the esophagus and is frequently associated with a diverticulum of Kommerell (4). Mirror-image branching can rarely occur, with the right aortic arch giving off, from left to right,

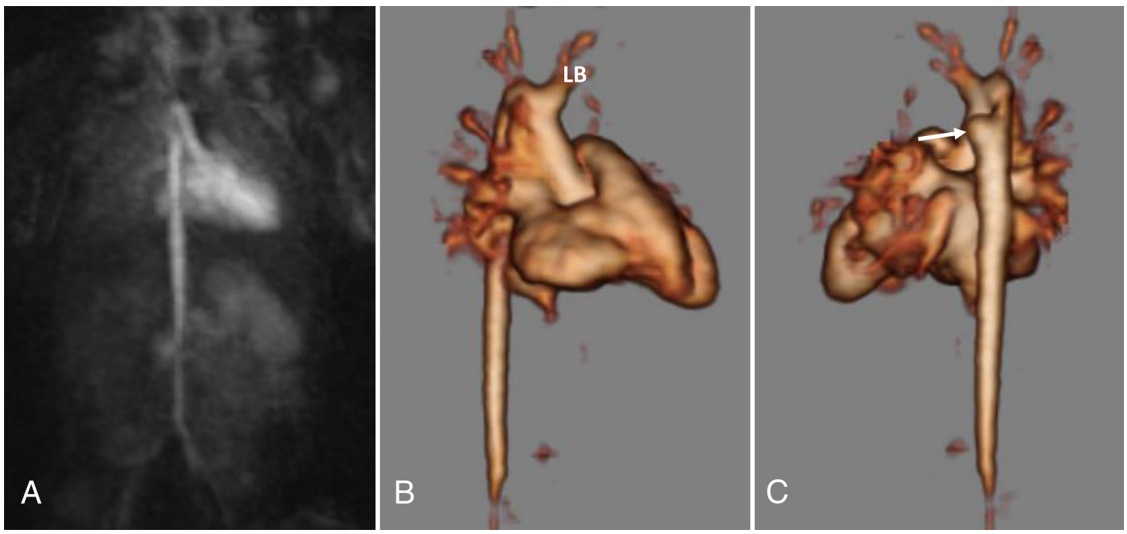

Figure 2. Dynamic contrast-enhanced magnetic resonance angiogram in a coronal projection showing contrast within the heart and the right aortic arch (A). Volume rendering shows a right aortic arch with mirror-image branching including a left brachiocephalic (LB) artery in an anterior view (B). Volume rendering shows a right aortic arch with a diverticulum of Kommerell (arrow) at the aortic isthmus in a posterior view (C).

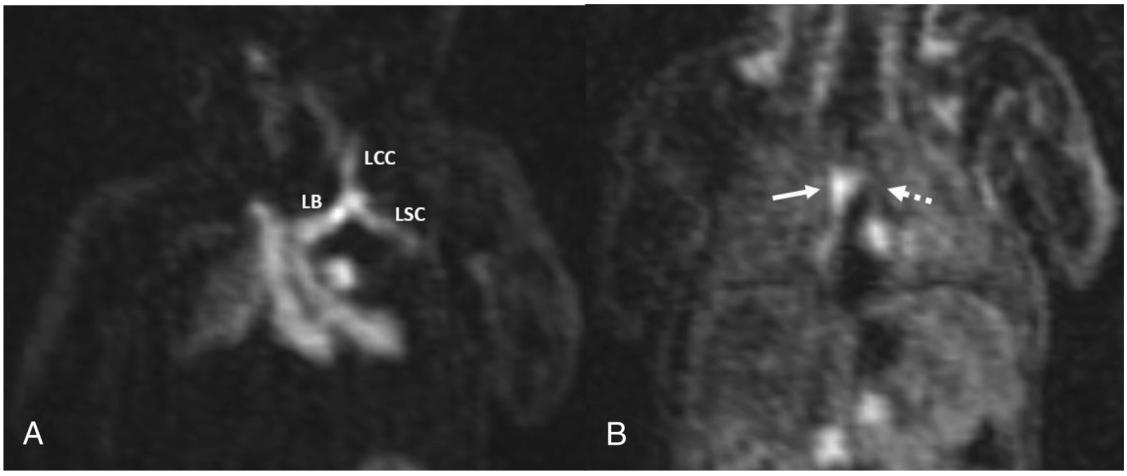

Figure 3. Dynamic contrast-enhanced magnetic resonance angiogram in an oblique projection shows the mirror-image branching pattern of the right aortic arch with a left brachiocephalic (LB) artery, left common carotid (LCC), and left subclavian (LSC) branches (A). 3D time-of-flight magnetic resonance angiogram in a coronal projection shows a diverticulum of Kommerell (arrow) at the aortic isthmus with a left ligamentum arteriosum (dashed arrow) posterior to the esophagus (B).

a left brachiocephalic artery (giving rise to the left common carotid and left subclavian artery), the right common carotid, and the right subclavian before continuing as the descending aorta (1).

The diverticulum in our patient at the aortic isthmus has been well described as the diverticulum of Kommerell, but it is only rarely seen with a mirror-image branching pattern (5). The patient also had an unusual ductus/ligamentum arteriosum originating from the diverticulum. The presence of a left ductus/ligamentum arteriosum between the aorta and the left pulmonary artery created a complete vascular ring structure comprising the ascending aorta anteriorly, the arch of aorta laterally, the descending aorta posteriorly, and the ductus/ligamentum arteriosum medially, which is capable of constricting the trachea and esophagus (6). Similar to our patient, in 74% of patients, the vascular rings cause symptoms including stridor and dysphagia, caused by the constriction of the trachea and esophagus, respectively (3). In these symptomatic patients, surgical division of the ductus or ligamentum can be performed to alleviate pressure on the trachea and esophagus (7).

This report shows the utility of neonatal cardiovascular MR imaging. Our patient initially underwent echocardiography with limited visualization of the aortic arch anatomy and left ligamentum arteriosum. The complex anatomy of the arch was clearly depicted on CMR despite the fact the infant was 3 days old and had no sedation. Cardiovascular MRI on neonates is performed ~2 times/month at our institution, and all of our neonatal studies for this indication are performed without sedation. This is possible until ~3–6 months of age. The entire procedure takes ~1 hour. As opposed to cardiac computed tomography (CT), cardiac MRI does not require any heart rate restriction to evaluate the cardiovascular structures. CMR also has the added advantage of defining the relationship of the vascular ring with the adjacent esophagus and trachea. Because of the high heart rates in neonates, CT of the aortic root is limited, premedication may be required, and evaluation of concomitant coronary or cardiac abnormalities may be limited. CT also poses the issue of ionizing radiation, and some centers do not have appropriate CT scanners to perform low-dose cardiac CTs on neonates. Furthermore, some locations lack radiologists or cardiologists with the expertise to read neonatal cardiac CTs. In contrast, limitations of cardiac MRI include increased cost, longer scan times, and higher levels of specialized training required to interpret the study. Additional investigation in a larger population is needed to determine the benefit of neonatal CMR in complex congenital arch abnormalities. Considering the widespread availability of CMR, this is certainly feasible.

Disclosures: No disclosures to report.

Conflict of Interest: The authors have no conflict of interest to declare.

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